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Case Report

Intranasal endoscopic surgery combined with adjuvant radiation therapy for olfactory neuroblastoma

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ABSTRACT

Olfactory neuroblastoma, also known as esthesioneuroblastoma, is a rare malignant head and neck tumor. Olfactory neuroblastoma accounts for approximately 3–5% of all malignant nasal tumors. We present a case of olfactory neuroblastoma in a 61-year-old man who underwent endoscopic surgery followed by postoperative irradiation. Right neck metastasis was diagnosed about 6 months after endoscopic surgery. Supraomohyoid dissection of the right side of the neck was performed, but right parotid metastasis was identified about 3 months later. Boost radiotherapy was applied to the right metastatic parotid. There was no evidence of recurrence on head and neck magnetic resonance imaging and endoscopic examination during the 2-year follow-up.

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1. Introduction

Olfactory neuroblastoma, also called esthesioneuroblastoma, is a rare head and neck malignant tumor. Olfactory neuroblastoma accounts for approximately 3–5% of all malignant nasal tumors [1]. Here we describe a patient with olfactory neuroblastoma who underwent endoscopic surgery followed by postoperative irradiation.

2. Case report

A 61-year-old man had experienced occasional nasal obstruction and epistaxis when sneezing for 2 years. Anosmia was also reported. Physical examination showed a bilateral nasal tumor that bled easily (Figs. 1A and 1B). Computed tomography revealed an infiltrative, enhancing mass lesion in the bilateral nasal meatuses and ethmoid and sphenoid sinuses, with invasion and destruction of the walls of the sinuses (Fig. 2A). The skull base was invaded by the tumor through the left cribriform plate (Fig. 2B). No neck or parotid metastasis was found (Figs. 2C and 2D).

We performed a biopsy under local anesthesia. Histopathological examination revealed clusters of infiltrating small round blue cells with hyperchromatic nuclei, a scanty delicate cytoplasm, and scattered mitoses (Fig. 3). The morphological picture was that of olfactory neuroblastoma. An immunohistochemical study revealed that most of the tumor cells reacted positively for the neuronal markers cytokeratin, synaptophysin, and chromogranin-A. Because the tumor had invaded the cribriform plate and skull base, it was classified as Stage C according to the Kadish classification (Table 1). The tumor was located over the bilateral nasal cavity and through the posterior portion of the nasal septum, and it bled easily on touch. Preoperative evaluation, including an abdominal ultrasound and whole-body bone scan, demonstrated no distant metastasis. Piecemeal excision of the intranasal tumor was performed carefully by a microdebrider. No serious complications, such as leakage of cerebrospinal fluid or intracranial hemorrhage, were observed in this patient. The patient underwent postoperative tumor bed radiotherapy of 6120 cGy for 7 weeks.

About 6 months after endoscopic surgery, a 3-cm × 3-cm mass was noted over the right neck Level II. Supraomohyoid neck dissection of the right side was performed, and pathological examination showed metastatic carcinoma similar to the previous olfactory neuroblastoma. A postoperative examination with whole-body ¹⁸F-fluorodeoxy-glucose positron emission tomography performed about 3 weeks after the procedure demonstrated no

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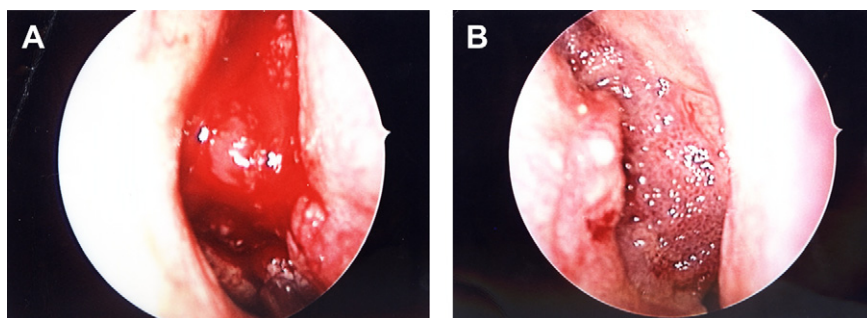


Fig. 1. (A) A nasal tumor that bleeds easily is observed on endoscopy of the right nasal cavity. (B) A nasal tumor is seen over the left nasal cavity through the posterior portion of the nasal septum.



Fig. 2. (A) Axial CT shows an infiltrative, enhancing mass lesion in the bilateral nasal meatuses with invasion and destruction of the sinus walls. (B) Coronal CT shows that the skull base has been invaded by the tumor. (C and D) No neck or parotid metastasis is seen. CT = computed tomography.

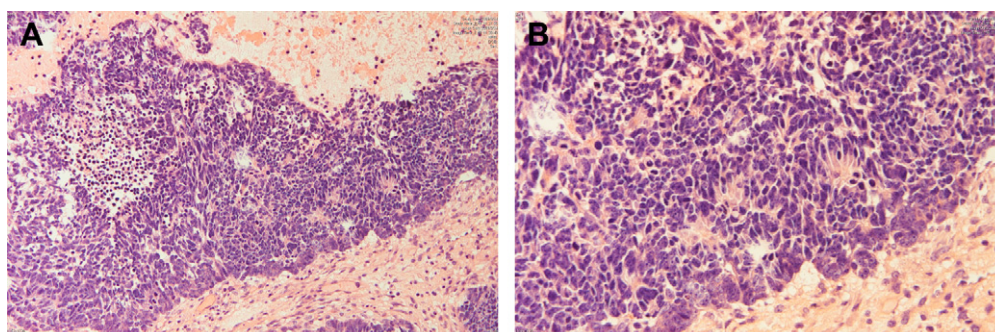


Fig. 3. (A) Microscopic view shows clusters of infiltrating small round blue cells with hyperchromatic nuclei, a scanty delicate cytoplasm, and scattered mitoses (H and E, original magnification: 200×) (B) Many rosette-like structures are seen (H and E, original magnification: 400×). H and E = hematoxylin-eosin stain.

Table 1
Kadish olfactory neuroblastoma staging.

Stage	Definition
A	Tumor limited to nasal cavity
B	Tumor confined to nasal and paranasal sinuses
C	Tumor extending to cribriform plate, base of skull, orbit, or intracranial cavity

significant ^{18}F -fluorodeoxy-glucose-avid hypermetabolic lesions in the nasal cavity (Fig. 4). However, right parotid swelling and tenderness were noted by the patient 5 weeks after the positron emission tomography. A right parotid sonography revealed an ovoid hypoechoic nodule about 1.5 cm \times 0.86 cm over the superficial lobe of the right parotid gland. A fine-needle biopsy was performed, and cytology revealed metastatic carcinoma. Because the patient was scheduled to receive radiotherapy, a total dose of 7200 cGy radiation was boosted to the right metastatic parotid. Head and neck magnetic resonance imaging and endoscopic examination showed no evidence of recurrence during the 2-year follow-up.

3. Discussion

Olfactory neuroblastoma is a locally extensive malignancy that frequently invades the skull base, orbits, and adjacent vital organs. Up to 70% of the patients were in advanced tumor stages at the time of initial diagnosis [2]. In Hyam's classification (Table 2), tumors are graded 1 through 4, with those showing less differentiation receiving higher grades [3]. The Kadish system is based on the spread of the tumor [4]. In this system, Stage A tumors are confined to the nasal cavity; Stage B tumors involve the paranasal sinuses; and Stage C lesions extend over the paranasal sinuses, including the

Table 2
Hyam's histopathological grading system of olfactory neuroblastoma.

Grade	Lobular architecture preservation	Mitotic index	Nuclear polymorphism	Fibrillary matrix	Rosettes	Necrosis
I	+	None	None	Prominent	HW rosettes	None
II	+	Low	Moderate	Present	HW rosettes	None
III	+/-	Moderate	Prominent	Low	FW rosettes	Rare
IV	+/-	High	Marked	Absent	None	Frequent

FW = Flexner-Wintersteiner; HW = Homer-Wright.

cribriform plate, base of the skull, orbit, or intracranial cavity. This patient had a Grade 3 Stage C lesion that had invaded the cribriform plate and skull base.

Surgery is the mainstream treatment for olfactory neuroblastoma. Traditionally, cranial facial resection entails serious complications, morbidity, and long hospitalizations [5]. The most common complications include leakage of cerebrospinal fluid, intracranial hemorrhage, and infection. In recent years, many surgeons and oncologists have begun treating olfactory neuroblastoma with a minimally invasive approach with good results [6–8]. Our patient was treated with combined intranasal endoscopic resection and radiotherapy. No other major complications were observed despite the relatively high incidence of complications in craniofacial series.

Distant and regional metastases are present in 14–38% of patients with olfactory neuroblastoma; the cervical lymph nodes, lung, and bone are the most commonly involved sites [9]. Our patient had right neck metastasis 6 months after the operation. The issue of elective treatment of the cervical lymph nodes in olfactory neuroblastoma is controversial. It is not clear whether prophylactic neck irradiation alters the natural history of the disease or adds a survival benefit [10].

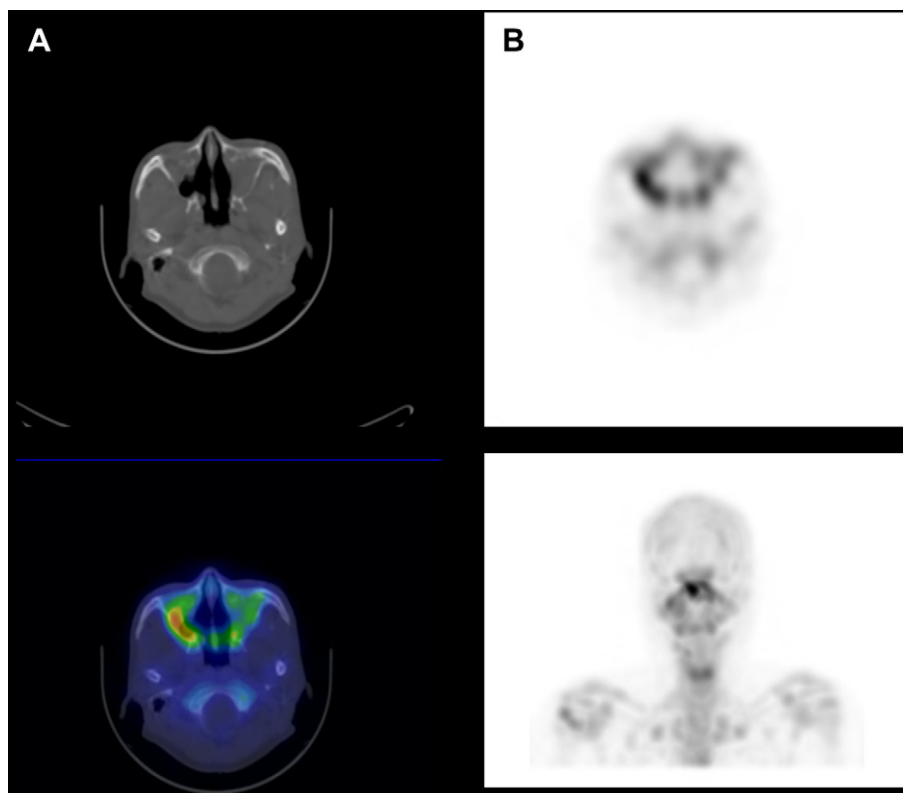


Fig. 4. Postoperative positron emission tomography scan performed about 3 weeks after the right neck dissection demonstrates no significant lesion in the bilateral parotids or neck.

Olfactory neuroblastoma is a very uncommon malignant tumor arising from the olfactory epithelium and is characterized by frequent local or regional recurrence. Combined intranasal endoscopic resection and radiotherapy minimizes injury to the patient. However, a longer follow-up is needed to determine the oncological outcome for this patient.

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